OPERATIONS PERFORMED AND VASCULAR ANOMALIES ENCOUNTERED IN THE TREATMENT OF CONGENITAL PULMONIC STENOSIS*

by

Alfred Blalock, M.D., Hon. F.R.C.S. and Henry T. Bahnson, M.D., Baltimore, Maryland

Department of Surgery of The Johns Hopkins University and The Johns Hopkins Hospital

AT THE PRESENT TIME there are three types of congenital heart disease that are amenable to surgical treatment. The first type to be treated successfully was the patent ductus arteriosus. This abnormality consists of the persistence into later life of the ductus arteriosus as a communication between the pulmonary artery and the aorta. There is no cyanosis, but there may be retarded physical growth, cardiac failure, or the development of endarteritis. The proper treatment, obviously, is interruption of this unnecessary and detrimental channel. Coarctation of the aorta is the second condition, which responds favourably to surgical therapy. Associated with the constriction in the aortic isthmus, these patients are prone to the development of hypertension, aneurysm and rupture of the aorta, and cerebrovascular accidents. In the majority of these patients the coarcted area of the aorta can be surgically excised and an end-to-end anastomosis performed with excellent results. The third group is made up of patients who are constantly cyanotic to a greater or lesser degree as a result of inadequate pulmonary blood flow and shunting of mixed venous blood to the arterial circulation. It is this condition, the commonest form of which is the tetralogy of Fallot, that forms the subject of this report.

Fig. 1A demonstrates the four anatomical features of the tetralogy of Fallot. These are: (1) Pulmonic stenosis or atresia, usually in the infundibular region of the pulmonary conus and occasionally involving the pulmonary valves, (2) a high interventricular septal defect situated in the membranous part of the septum beneath the origin of the great vessels, (3) a dextroposed aorta overriding the septal defect, and (4) hypertrophy of the right ventricle. As a result of the shunting of mixed venous blood through the overriding aorta, patients with this condition constantly have some unsaturation of the arterial blood, which increases when bodily demands are greater, as upon exertion. An inadequate blood flow to the lungs accompanies the pulmonic stenosis, and only a fraction of the venous blood entering the right auricle courses through the lungs for oxygenation. The intolerance to exercise and

*Moynihan Lecture delivered before the Royal College of Surgeons, September 26, 1947, by Alfred Blalock.

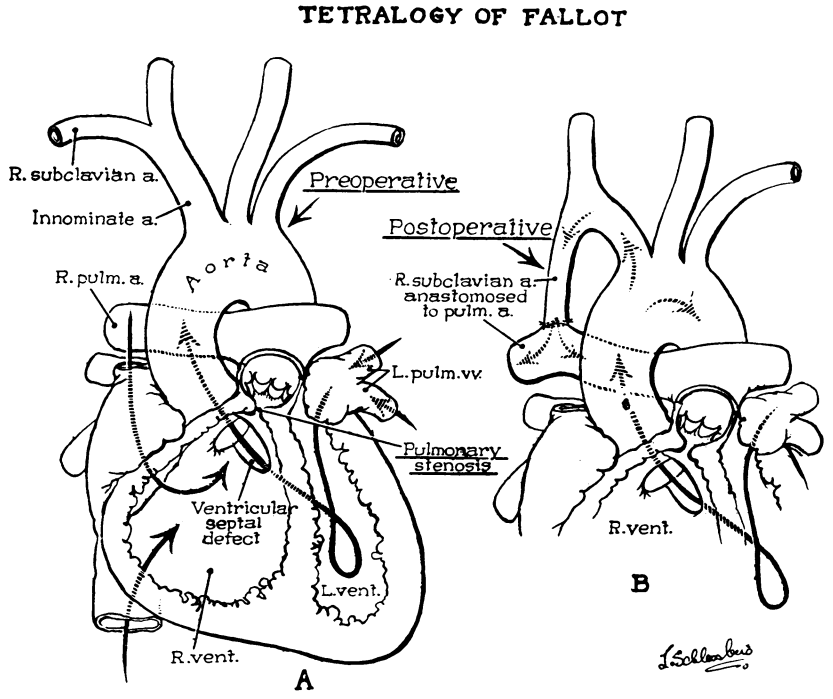

Fig. 1A. Diagram showing the tetralogy of Fallot. This demonstrates the pulmonic stenosis in the conus region, the interventricular septal defect, the aorta which overrides the defect and receives some venous blood, and the right venticular hypertrophy.

Fig. 1B. Diagram showing the tetralogy of Fallot after operation. The proximal end of the right subclavian artery has been anastomosed to the side of the right pulmonary artery, thus shunting incompletely oxygenated blood from the aorta into the pulmonary arteries.

the cyanosis are present because of these two components of the tetralogy—namely, the overriding aorta and pulmonic stenosis. It is only because of the coexistence of these two factors that successful surgical therapy can be carried out. It is apparent that if some of the improperly oxygenated blood from the aorta can be shunted through the pulmonary arteries by an artificially created ductus arteriosus, it will acquire more oxygen without overloading the diminished pulmonary circulation. Fig. 1B shows the arrangement of the circulation following the completion of an end-to-side anastomosis. In contrast to this condition is the one known as pure pulmonic stenosis, a rare abnormality in which there is no septal defect or overriding aorta. All of the blood returning to the left side of the heart and subsequently being ejected through the aorta is properly oxygenated and hence, although the volume of pulmonary blood flow may be greatly reduced, a great deal cannot be gained by shunting aortic blood through the lungs again.

There are, however, many congenital cardiac conditions which are amenable to the creation of an artificial ductus arteriosus. These include the tetralogy of Fallot with pulmonic stenosis or atresia, a single ventricle with pulmonic stenosis, tricuspid and pulmonary atresia, pulmonic stenosis with an interauricular septal defect, transposition of the great vessels with pulmonic stenosis and a large interventricular septal defect, and many variations of these abnormalities. Perhaps patients with an interventricular septal defect and an overriding aorta but without pulmonic stenosis, as in the Eisenmenger's complex, may be benefited, although this point has not been determined. Certainly the primary indication for operation is an inadequate blood flow to the lungs with an interventricular septal defect and an overriding aorta.

The criteria for the diagnosis of pulmonic stenosis or atresia have been described in detail by Dr. Taussig (2) and others. Cyanosis, dyspnea, poor tolerance for exercise, squatting, a systolic murmur in the pulmonary area, low arterial oxygen saturation, polycythemia, a concavity in the region of the pulmonary artery on X-ray examination, a clear pulmonary window, diminished hilar shadows, and absent pulsations at the lung hila on fluoroscopic examination, are the more important positive points in diagnosis. In general, the most important information is obtained by roentgenography and fluoroscopy. Dr. Richard Bing and his co-workers (3) have described a valuable laboratory test which consists of measuring the ratio of oxygen consumption to volume of ventilation before and after exercise. In normal persons this ratio increases, in contrast to those with an inadequate pulmonary blood flow in whom the ratio falls.

Technique of Operation

The possibilities of operative treatment of the tetralogy of Fallot and allied conditions include a direct attack upon the pulmonic stenosis and

the performance of a shunting operation by which the stenotic area is by-passed. In contrast to acquired heart disease, congenital pulmonic stenosis is usually below the valves and involves the pulmonary conus. An incision into this region would not only be dangerous, but the effects would probably be only temporary as further scarring occurred. The performance of a shunt operation, on the contrary, is facilitated by the proximity of the aortic arch and its branches to the pulmonary arteries. Because of the large pressure gradient between these vessels, we may expect an anastomosis to remain patent and to carry a large volume of blood into the pulmonary circulation.

There are a number of procedures by which the systemic and pulmonary circulations may be connected in the performance of the shunt operation. The subclavian branch of the innominate artery may be anastomosed to the end or to the side of the pulmonary artery to the lung on that side of the body. The carotid or the innominate artery may be similarly anastomosed. The subclavian artery which arises directly from the aorta may be anastomosed to the end or the side of one of the pulmonary arteries, or lastly, the aorta may be anastomosed directly to one of the major branches of the pulmonary artery in a side-to-side manner. The type of operation must be selected to fit the individual case. We prefer to use the subclavian artery, which arises from the innominate and to make an end-to-side anastomosis, because the subclavian artery arising from the innominate makes a better angle after completion of the anastomosis than that which arises directly from the aorta. An end-to-side procedure is preferred because it allows the shunted blood to go to both lungs with less danger of an excessive burden on either.

In order to utilize the subclavian artery one must determine preoperatively the position of the aortic arch, since in a large number of cases in this group of patients the aorta arches over the hilum of the right lung instead of the left. The radiological method described by Bedford and Parkinson[4] is used to ascertain the side. The innominate artery remains the first vessel to arise from the aorta, regardless of whether the arch is to the right or the left. Hence, if one wishes to use the subclavian artery arising from the innominate, it can best be approached from the right with a left aortic arch or from the left with a right aortic arch.

The various steps in the operation which we usually perform may be seen in Figs. 2 to 7. The patient is placed on his back with the operative side slightly elevated and on a table which may later be tilted away from the operator. The incision is made from the edge of the sternum to the apex of the axilla. In females it is thought advisable to circle beneath the breast in making the skin incision. In the majority of the patients the chest was entered through the third intercostal space. It is now our policy to utilize the second interspace in all patients except infants, since this allows a more complete expansion of the lung on the operated side

as well as a better exposure of the apex of the chest. The azygos vein is divided, a step which permits freer dissection in the hilum of the lung and exposure of the pulmonary artery. This vessel is mobilized as completely as possible. Dissection is then carried beneath the superior vena cava, and the innominate artery is identified as it arises from the aorta. In following this vessel distally to its branches one may be guided by the vagus and recurrent nerves in locating the subclavian artery. After these structures are identified it is possible to divide the mediastinal tissue with greater freedom. It is important to free the subclavian, carotid and innominate arteries in order to gain as much mobility as possible for the vessel to be anastomosed. A rubber-shod clamp is applied to the subclavian artery proximally. A ligature is placed distally where the artery branches, and it is divided just proximal to the ligature. The end of the subclavian artery is freed of adventitia to avoid its being caught in the suture and causing constriction of the anastomosis. At this point the lung is inflated and the patient is allowed to rest.

The pulmonary artery is occluded proximally with a special clamp and distally with rubber-shod clamps and traction upon untied silk ligatures which have been placed around the branches. A transverse opening is made in the superior surface of the pulmonary artery a trifle larger than the end of the subclavian. Again the adventitia is removed to avoid its inclusion in the suture line. An anastomosis is performed between the end of the subclavian and the side of the pulmonary artery with 5-0 silk on an atraumatic needle. The suture is a continuous one, everting and approximating the intima. The posterior row is placed before it is pulled taut, and additional stay sutures are placed at either end to anchor the continuous suture. The anterior row is completed by using a similar stitch. This part of the anastomosis is usually easier than placing the posterior row. There is often a slight ooze from the suture line on removal of the occluding devices, but this usually stops with brief pressure. If there is a considerable leak it can be stopped with an additional suture.

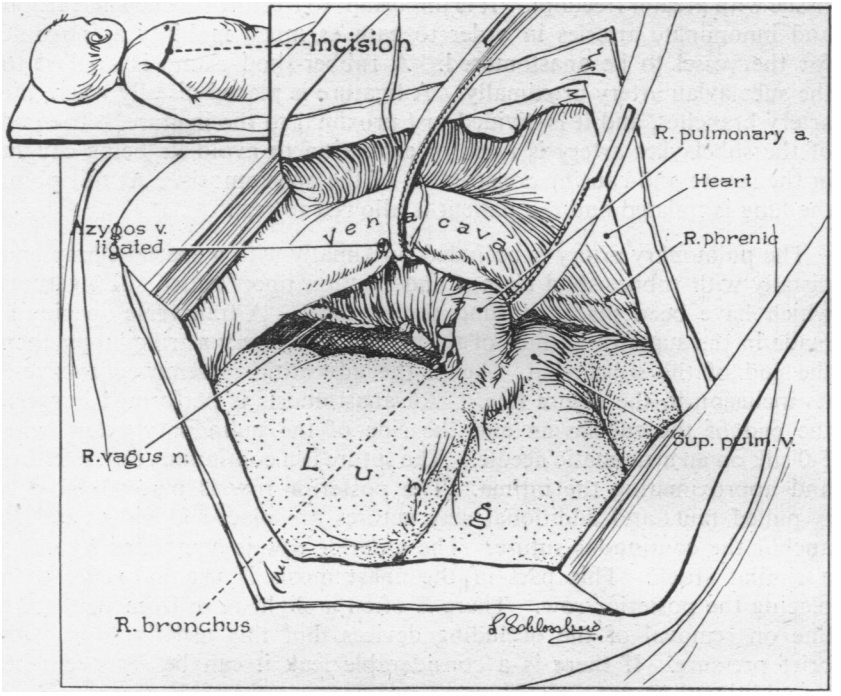

Fig. 2. Inset shows the position of the patient for a right thoracotomy. The azygos vein has been divided and the superior vena cava is retracted with it. The right pulmonary artery has been freed and a clamp placed under it. The artery should be thoroughly mobilized and the branches freed laterally.

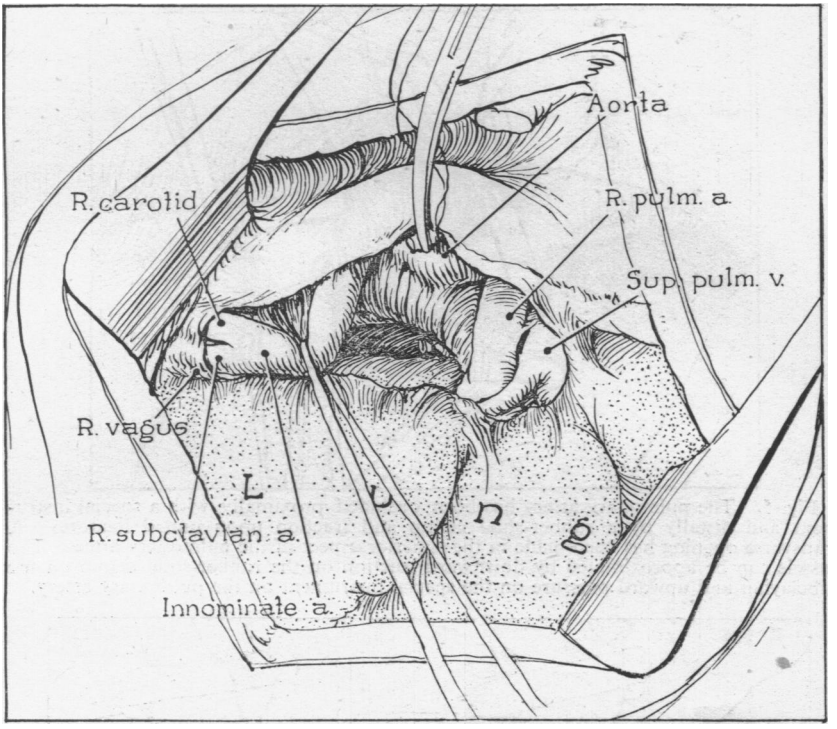

Fig. 3. By dissection under the superior vena cava the innominate artery has been identified and freed proximally to the aorta and distally to the branches. The vagus nerve is seen.

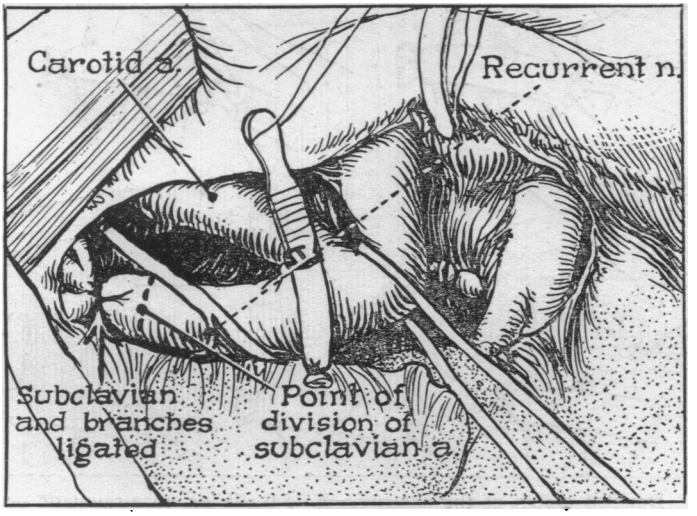

Fig. 4. The right subclavian artery has been freed and ligatures have been tied around the branches. A rubber-shod clamp has been placed on the artery near its origin and tied in place to prevent slipping. The artery will be divided just proximal to the ligature.

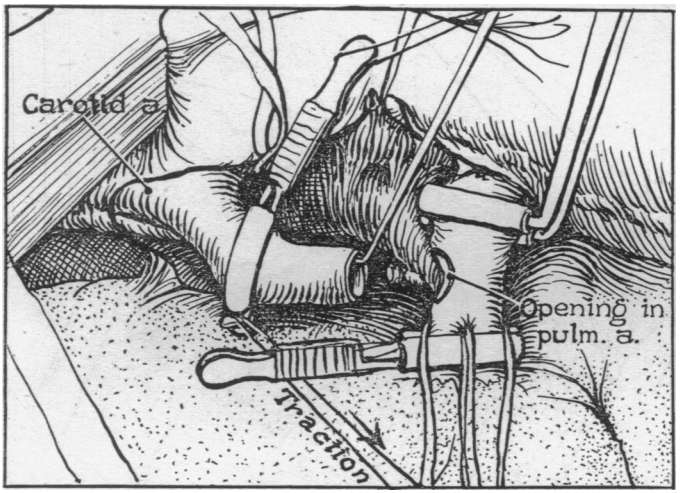

Fig. 5. The pulmonary artery has been occluded proximally with a special instrument and distally by a rubber-shod clamp and traction upon untied ligatures. A transverse opening has been made in the superior aspect of the pulmonary artery. The vessels can be approximated by downward traction on the rubber-shod clamp on the subclavian and upward pressure on the special instrument on the pulmonary artery.

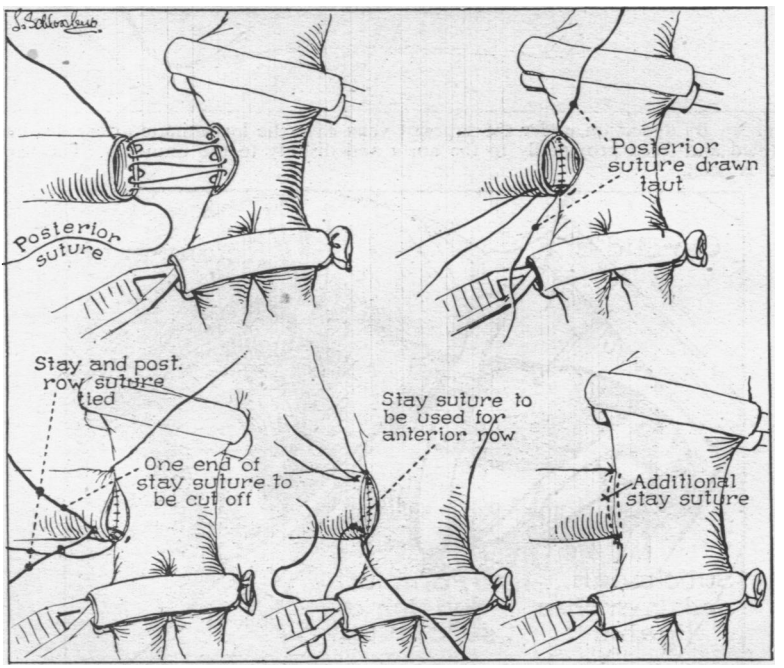

Fig. 6. Detailed sketch of the anastomosis. 5-0 silk on an atraumatic needle is used. The suture is continuous, everts the wall of the vessel, and approximates the intima. The posterior row is placed before it is drawn taut. Actually very little silk shows after the suture is drawn taut. The interrupted sutures are placed for anchorage and to prevent a purse-string effect.

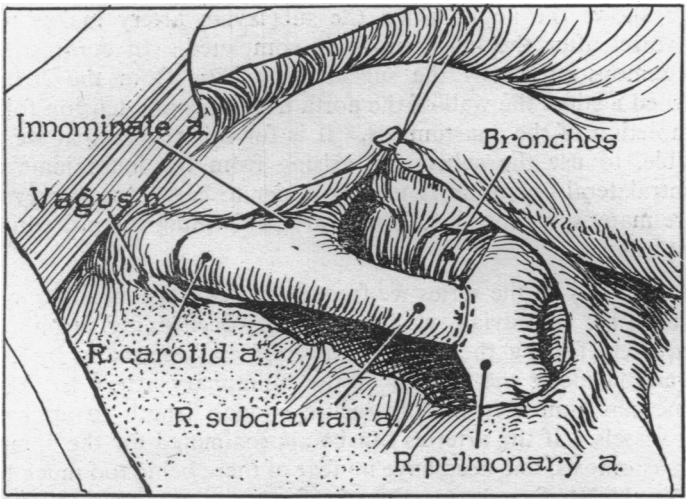

Fig. 7. The end-to-side anastomosis has been completed. Note the angle which the subclavian artery makes with its parent vessel, the innominate. Compare this with Figure 8.

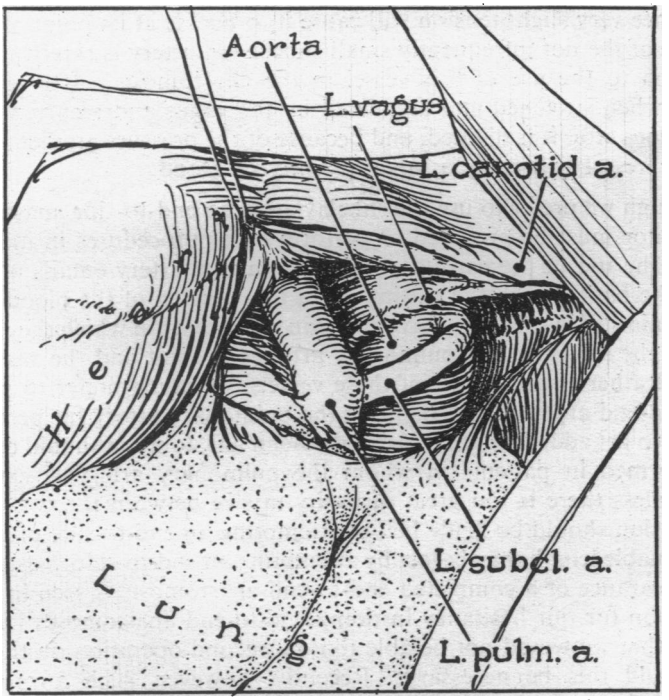

Fig. 8. An end-to-side anastomosis made by using the subclavian artery which arises from the aorta. Note the angulation at the origin of the artery. In some cases there may be enough tension actually to flatten the subclavian. This may be lessened by suturing the pleura over the lung to the mediastinal pleura and by inflating the lung.

Fig. 7 shows the angle which the subclavian artery makes with its parent vessel after the anastomosis is completed. In contrast to this Fig. 8 demonstrates how the subclavian arising from the aorta may be flattened against the wall of the aorta if there is any tension following the completion of the anastomosis. It is for this reason that we prefer, if possible, to use the subclavian arising from the innominate artery. The contralateral operation is usually easier to carry out, however, and there are many surgeons ([5], [6], [7]) who are performing the procedure with excellent results.

Although early in the series we frequently made use of other systemic vessels than the subclavian, we have recently been doing so only rarely. By adequately freeing the systemic arteries and especially by dividing the adventitia along the branch of the pulmonary artery to the lower lobe, one can almost invariably secure enough length to approximate the two vessels. If the arteries can be approximated for the completion of the anastomosis, one need have no fear of there being too much tension when doing the preferred operation, for further approximation is attained by the removal of the occluding devices and the inflation of the lung. This is not the case when one uses the subclavian arising directly from the aorta since very slight tension will cause it to buckle at its point of origin. The size of the not infrequently small subclavian artery is rarely a contra-indication to the use of this vessel in the anastomosis. It often looks smaller when stretched and angulated by the vagus and recurrent nerves than it does after it is divided, and because of the pressure gradient a small systemic vessel will carry a large quantity of blood.

Although we prefer to use the subclavian in an end-to-side anastomosis, an operator must be prepared to perform other procedures in individual cases. The use of the carotid or the innominate artery entails a greater risk to the patient with the cutting off of this portion of the blood supply to the brain. This may be unavoidable in some cases in which the distance between the systemic and pulmonary arteries is great and the vessels are short. Rather than use one of these vessels, we much prefer to perform an end-to-end anastomosis, dividing the pulmonary artery far proximally in order to get added length. An end-to-end anastomosis should certainly be performed in patients in whom the pulmonary artery is extremely small, unless there is too great disparity in size between the two vessels. The decision should be made before attempting an end-to-side procedure, since valuable length may be lost by attempting an end-to-side anastomosis. The appearance of a completed end-to-end anastomosis is seen in Fig. 9. One reason for our hesitancy in doing end-to-end anastomoses has been the fear that we would not be able to do a second operation on the other side should this be necessary. Recently, however, such a procedure was carried out on a child who satisfactorily tolerated occlusion of the pulmonary artery and the remainder of the operative procedure with a dramatic result.

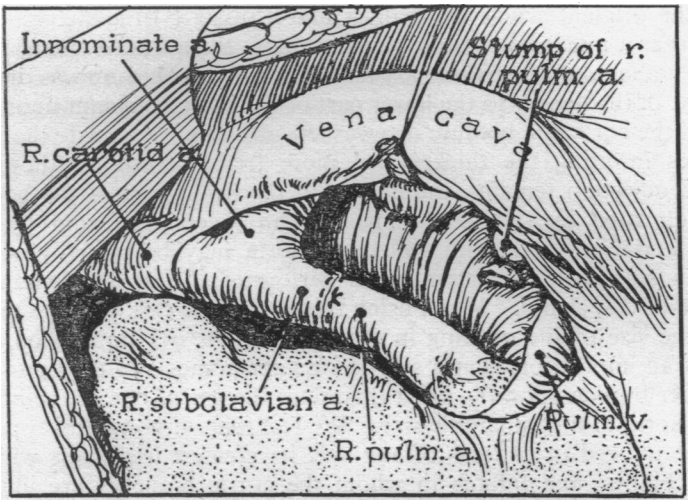

Fig. 9. Anastomosis between the end of the right subclavian and the end of the pulmonary artery because of a short subclavian. A modified Carrel technique is used, which consists of placing three guy sutures around the circumference of the arteries and suturing between them with an everting stitch.

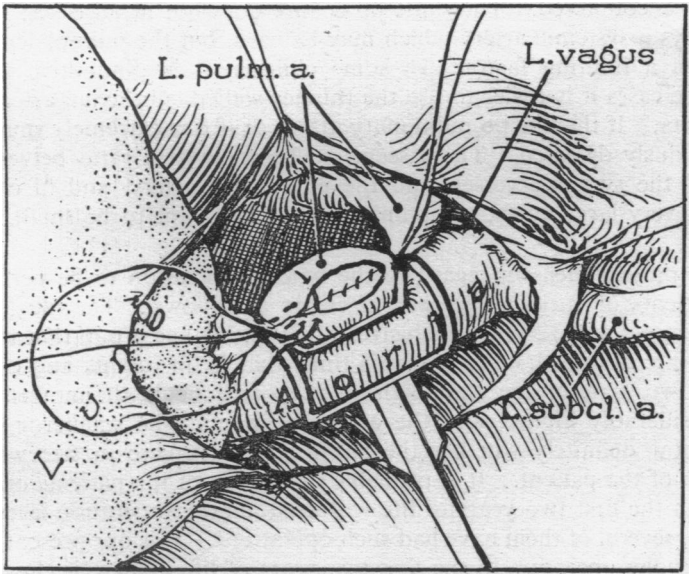

Fig. 10. Showing the operation as described by Potts, Smith and Gibson. The partial occlusion clamp is placed on the aorta and a side-to-side anastomosis is performed to the left pulmonary artery. The suture is pulled up as it is placed. The intima cannot be everted as well as when the thinner walled branches of the aorta are used.

In the original article by Blalock and Taussig (1) the suggestion was made that it might be possible to anastomose the aorta to one of the two major branches of the pulmonary artery, but this procedure was discarded because of the danger to the lower part of the body attendant upon occlusion of the aorta and because one was less able to approximate the intimal surfaces in using the thicker and more friable aorta. Although the second objection remains, the danger to the lower part of the body has been obviated by the development of an ingenious clamp by Potts, Smith and Gibson(8), with which the aorta may be partially occluded while a side-to-side anastomosis is performed. Fig. 10 demonstrates the use of this clamp. When using this procedure care must be taken to limit the size of the opening in the aorta. It is our impression, and it is only an impression, that a side-to-side anastomosis of the aorta places a greater burden upon the heart than does the use of one of the branches of the aorta of a comparable size. We feel that, although this operation may be utilized in patients of any age, its greatest usefulness will be in small children and infants in whom the aortic branches are unusually small.

The principle underlying all of these variants is the same—namely, the by-passing of the point of obstruction to the flow of blood to the lung. The surgeon performing this type of work must be prepared to do any of the operations mentioned according to the indications and the conditions encountered. The same procedure does not fit all cases. There is always a systemic artery which may be used, but the pulmonary artery is often a limiting factor. In some children it is diminutive in size. In these cases it is easier to use the thinner walled subclavian artery than the aorta. If there is no pulmonary artery or if it is extremely small, one is obviously defeated. There seems to be an inverse ratio between the size of the systemic vessels and the pulmonary artery, and in children with a very large aorta, the pulmonary artery is apt to be small.

Our present ideas in regard to the type of operation to be performed on patients of various ages and sizes are as follows: The ideal age is between two and 12 years, inclusive. It is in this age group that the best results are obtained. Prior to this time, that is, in infants and children under two years of age, the diagnosis is more difficult, the mortality rate is considerably higher, and the anastomosis must be smaller. We do not know definitely whether the anastomoses increase in size with the growth of the patient. It is probable that many of the patients operated upon in the first two years of life will need another operation later, and, indeed, several of them have had such operations. It is our present policy to postpone operation in the first two years of life if there is thought to be a 50 per cent. chance that the patient will survive until his second birthday. If operation is decided upon, we operate on the side on which the aorta descends and use the subclavian artery if it is large enough; if

it is not, we perform a side-to-side anastomosis with the aorta. In patients in the 2-to 12-year age group we operate upon the side opposite to that of the aortic arch and prefer to use the subclavian branch of the innominate in an end-to-side anastomosis. If this is impractical because of too great a distance between the systemic and pulmonary vessels or because of small pulmonary arteries, an end-to-end anastomosis is performed. The carotid and innominate arteries are used with great reluctance.

We know of no specific way whereby one may render patients older than 12 years more suitable for operation. In these older patients the mortality rate is slightly higher and the operation is more difficult because of the increase in the collateral vessels in the lung hilum and mediastinum, and the decreased elasticity of the arteries and the increased distance between the systemic and pulmonary arteries. In these patients and in younger ones who are more than five feet in height we make the incision on the left. When the aortic arch is on the left the subclavian artery is used, if possible; if this is impossible, a side-to-side anastomosis is performed between the pulmonary artery and the aorta. If the aorta descends on the right, the subclavian branch of the innominate is used. This vessel is usually a long one and, as stated previously, the left pulmonary artery is higher than the right so that approximation is easier. Fig. 11 shows such an anastomosis completed.

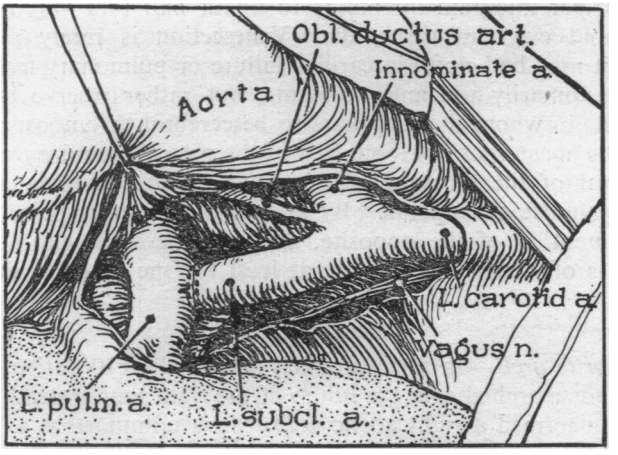

Fig. 11. Anastomosis of the left subclavian to the left pulmonary artery in a patient with a right aortic arch. Note the obliterated ductus arteriosus joining the pulmonary artery to the distal innominate. The operation is usually easier on this side because the left pulmonary artery is higher than the right.

The pre- and post-operative care of these patients is not complicated, but they do require frequent supervision, since a short time may mark a considerable change in their condition. Digitalis is given pre-operatively to the occasional patient with signs of decompensation. Venesections are not performed preparatory to operation. The fluid intake must be maintained to avoid dehydration and cerebral thrombosis. A child should receive at least 1,600 cc. of fluid a day and an adult at least 2,200 cc. The premedication is 1 mgm. of morphine per 10 pounds of body weight with 1/20th of this dose of atropine. In the majority of instances the anæsthetic is intratracheal cyclopropane and oxygen. In those patients in whom there is arrhythmia this is changed to ether and oxygen. The anæsthesia is as light as is consistent with a quiet mediastinum. Fluids are given in a quantity to match the estimated loss. Blood plasma is used unless the patient does not have polycythemia, in which case whole blood may be indicated. Any excessive loss of blood is replaced with whole blood. There are not infrequently short periods of bradycardia during the course of the dissection; although a cause for concern, these usually respond to the administration of intravenous atropine.

Post-operatively, oxygen tents are used routinely for a day or so, depending on the patient's condition. Prophylactic doses of penicillin are administered. The intravenous drip begun at operation is continued until the patient is taking adequate fluids by mouth. The fluid intake is restricted slightly in an effort to prevent overloading the pulmonary circulation, but at the same time dehydration with its attendant cerebral thrombosis must be avoided. The usual volumes range from 80 cc. to 100 cc. per kilogram in infants to about half that volume in older children and even less in adults. Venesection is rarely resorted to, although it may be helpful in cardiac failure or pulmonary œdema. We do not customarily use anticoagulants, but rather reserve heparin for the patients in whom there appears to be cerebral thrombosis or thrombosis of the anastomosis. About half of the patients require thoracentesis for removal of the serosanguineous fluid which accumulates to some degree in almost all patients. By and large one must be as careful of over-treatment as of the opposite, and rest, perhaps with the help of sedation, is of considerable value, at least for the first few days.

Vascular Anomalies

Right aortic arch.—In approximately 140 of the 610 patients operated upon the aorta arched over the hilum of the right lung instead of the left, a condition termed a right aortic arch. It is unimportant as far as the patient's symptomatology is concerned but must be ascertained before operation in order that one may enter the correct pleural cavity. It should be remembered that the innominate is the first branch of the aortic arch and can be best reached through the plural cavity opposite that of the aortic arch. Almost invariably the operation is easier when

performed with a right aortic arch, for the pulmonary artery is a few centimetres higher on the left than on the right, and the left subclavian in such cases is usually a longer vessel. In about three-fourths of the patients with a right aortic arch an obliterated ductus arteriosus is found coursing between the left pulmonary artery and the left subclavian artery or the innominate at its bifurcation (Fig. 11). However, the ductus is not always on the right in such patients, and, in fact, Dr. John Jones has operated on the right for a patent ductus in a patient with a right aortic arch.

The usual course of the aorta distal to the arch is a continuation of the thoracic aorta to the right of the œsophagus. In some instances, however, the aorta runs behind the œsophagus and enters the left chest cavity for the remainder of its intrathoracic portion. This is termed a right aortic arch with a left descending aorta. One patient was recently operated upon who had the rare combination of a left aortic arch, a retro-œsophageal aorta, and a right descending aorta.

Retro-œsophageal subclavian artery.—In 26 patients, 12 of whom were operated upon on the right side, the subclavian artery to the extremity on the side opposite the aortic arch arose as the last branch of the arch. It coursed behind the œsophagus and trachea to enter the opposite pleural cavity. In most instances this condition is suspected by the presence of a persistent filling defect in the posterior wall of the œsophagus demonstrated by barium swallow and fluoroscopy. This arrangement offers no contraindication to the use of this vessel, and, indeed, it is often of greater length and is more easily approximated to the pulmonary artery than the normal subclavian branch of the innominate artery. In one patient in whom a retro-œsophageal subclavian artery was anastomosed to the pulmonary artery, there were suggestive signs of tracheal obstruction post-operatively. The child died as a result of cerebral complications, and at autopsy there seemed to be some œsophageal and tracheal compression by the " vascular ring " created by the anastomosis. Since that time we have been freeing these vessels and transposing them anterior to the bronchus on the operated side before performing the anastomosis. For some unknown reason the retro-œsophageal arteries are thinner walled than normal arteries and must be handled with greater care.

In one case the innominate artery arose from the distal part of the aortic arch and was retro-œsophageal in its course prior to giving off its branches.

Absence of the innominate artery.—Abnormal arrangements of the origin of the great vessels arising from the aortic arch occur so frequently that, like a right aortic arch, they can hardly be called anomalies. All four vessels may arise from the arch independently, or one subclavian and both carotid arteries may come from a single trunk. A thyroid ima

artery may arise from the aortic arch, the innominate, or the carotid artery. In the latter two instances this is of some importance since it may be torn in freeing these vessels. Whenever one doubts the identity of systemic vessels, help is obtained by occluding them and having the anæsthetist feel the various pulses. In several instances we have failed to appreciate the presence of a retro-œsophageal subclavian artery and have used the carotid.

In one patient it was noted pre-operatively that the left carotid pulsation was weaker than the right. At operation on the left side with a right aortic arch no innominate artery was found. In its place a fibrous band ran from the left pulmonary artery to the site of the normal bifurcation of the innominate. The collateral circulation was such that vigorous back pulsations occurred on occlusion of either the carotid or the subclavian artery, and the child was improved following anastomosis of the side of this bifurcation to the side of the left pulmonary artery.

Double aortic arch.—In rare instances the right aortic arch may undergo its full development without disappearance of the left aortic arch. Under these circumstances there may be tracheal or œsophageal obstruction in some patients. The origin of the great vessels from the two arches is variable, but from the standpoint of embryology it would be expected that at least the four main vessels would arise independently. It may be possible to diagnose the condition before operation by fluoroscopic evidence of œsophageal compression from both of the arches. We are aware of only one patient in this series in whom there was a double aortic arch associated with a tetralogy of Fallot. The patient did well in her early post-operative period, but died a fortnight later. At autopsy a large ball thrombus was found in the left ventricle, and it was felt that this had suddenly occluded the left ventricular outlet.

Single pulmonary artery.—In nine patients it was demonstrated at operation or autopsy that there was no branch of the pulmonary artery to one of the lungs, the diminished function of the lung being maintained by bronchial and collateral vessels. This condition can be suspected at the time of operation by the failure of the patient to tolerate occlusion of the pulmonary artery or by the rise in pressure of the proximal segment when it is occluded distally. Of course, when one operates on the opposite side no pulmonary artery will be found. If one is aware of the condition, it might be possible to occlude the artery partially, much as the aorta is occluded in the Potts operation, but it is probable that the risk would be too great to warrant such a procedure.

Functional truncus arteriosus with a blind pulmonary artery.—There have been several patients in whom there was no connection between the pulmonary arteries and the heart, except through collateral and bronchial arteries. In one of these the right pulmonary artery at its bifurcation

and distally was of normal size and configuration, but only a fibrous strand existed to represent the proximal portion of the pulmonary artery. The pulmonary circulation was effected by a large retro-œsophageal artery, which entered the posterior aspect of the hilum of the lung. An anastomosis was begun to the side of the bifurcation of the pulmonary artery, but the child succumbed as this was being completed.

Bilateral superior venæ cavæ.—This condition is probably not rare, and is now being demonstrated with unexpected frequency by the greater employment of angiocardiography in puzzling cases. This arrangement of the venous return is, of course, not apparent at the time of operation, although persistent left venæ cavæ have been seen fairly often. When a left vena cava persists it usually enters the right auricle through the coronary sinus, since the cava and the sinus are the remnants of the left duct of Cuvier. In one patient, however, it was shown by angiocardiography that a right superior vena cava entered the right auricle and a left superior vena cava entered the left auricle. No treatment has been directed at the anomalous venous return, and in no case has it seemed to be of significance in causing cyanosis.

Other anomalies of the systemic veins.—In several patients in whom a left thoracotomy was performed, both superior and inferior venæ cavæ were encountered. The latter arched behind and over the hilum of the left lung to join the superior cava. The site of opening into the heart was not determined. In creating an artificial ductus arteriosus in such patients care must be taken not to constrict either of these vessels. The anastomosis is best constructed so that after completion it will lie medial to the vessels.

In one patient the left innominate vein was posterior to the innominate artery before joining the right innominate vein to form the superior vena cava.

Anomalies of the pulmonary veins.—One frequently sees pulmonary veins emptying into the right auricle, or the right, or a persistent left superior vena cava. The surgical significance of these in pulmonary surgery of patients without congenital heart disease has been shown by Brody (9), Brantigan (10) and others. Such anomalies are more frequent in patients with congenital defects. It has never seemed advisable to ligate these veins or to attempt to anastomose them to other structures.

Results

A total of 610 cyanotic patients have been operated upon in whom a pre-operative diagnosis of an inadequate pulmonary blood flow had been made by Dr. Taussig and her associates, Dr. Bing, with his physiological methods, and others. With the exception of 15 operations performed at Guy's Hospital, London, and Hôpital Broussais in Paris, these have all been done at The Johns Hopkins Hospital by members of the surgical

staff.* A second operation has been performed in several of these patients. In this total number there have been 108 deaths, an overall mortality rate of 17·7 per cent. No known deaths are excluded. Twenty-seven patients died at the time of operation, 68 while in the hospital during the post-operative period, and 13 after leaving the hospital. In a few of these patients only an exploration was performed as the pulmonary artery was not found or was not suitable for an anastomosis. In some the diagnosis was in error; for example, there were seven fatal cases where there was transposition of the aorta and pulmonary artery without pulmonic stenosis. A few of the deaths were attributable to unrelated causes such as meningitis, pneumonia, and so forth, after discharge from the hospital.

Of the 27 patients who died during the operation, 12 died in the course of exploration alone, either because the pulmonary artery was inadequate for an anastomosis or because of arrhythmia before the anastomosis could be attempted. We are not sure of the cause of death in these patients, and there probably is no single cause, but cardiac and cerebral anoxia seems to play the most important role.

Of the 68 patients who died in the hospital after operation, 24 died of cerebral complications, including cerebral thrombosis, anoxia, or brain abscess in a few; 16 died of cardiac failure or pulmonary œdema; hæmorrhage was the primary cause of death in eight, and thrombosis of the anastomosis in an equal number; three died with respiratory complications.

There have been 13 patients who died after leaving the hospital. Some of the deaths were not related to the operative procedure. The causes were varied and included pneumonia, cardiac failure, meningitis, cerebrovascular accidents, and coronary occlusion.

The mortality rate depends to a considerable extent upon the principles which one adopts in regard to the indications for operation. We have felt that any patient should be operated upon in whom available methods point to the diagnosis of inadequate pulmonary circulation. This has included many patients who were extremely poor operative risks, and, in fact, there have been a number of patients who died while in the hospital awaiting operation. Some of these extremely poor-risk patients, however, had good results following the completion of the operation. One's mortality rate will be lower if he selects only those patients who are in the age group of two to 12 years, without extreme incapacity or a history of cerebral accidents, with a small heart, a good systolic murmur,

* This includes Doctors Longmire, Shumacker, Scott, Hanlon, Bahnson, Duncan, Clay and Cantrell, who performed a total of 149 operations.

and the other features of a typical tetralogy of Fallot. On the other hand, the number of deaths will obviously increase if indications are extended to include patients with a rotated heart, arrhythmia, cardiac enlargement, evidence of cerebral damage, and pulmonary atresia as suggested by the absence of a systolic murmur, and infants and older adults. In selected cases in which one is able to perform the preferred anastomosis the deaths are fewer. Of the 433 patients in whom a subclavian-pulmonary anastomosis was performed end-to-side, 10·4 per cent. died, and in a few of these there was an error in diagnosis. In one sequence we operated upon 44 consecutive patients without a death.

The end of the subclavian artery has been anastomosed to the end of the pulmonary artery in 38 cases, with 15·7 per cent. mortality In the majority of these patients the indication for the end-to-end anastomosis was a small pulmonary artery. Contrary to opinions held at the beginning of this work, the use of the innominate or carotid artery for an anastomosis is seldom necessary. The use of the innominate artery in 49 patients was associated with a 30·6 per cent. mortality and of the carotid in 34 patients with a 23·5 per cent. mortality. Admittedly these patients are often infants or very sick children who are poor risks, but the use of these vessels adds to the danger of the operation. The aorta has been anastomosed to the left pulmonary artery in two patients, one of whom died as a result of pulmonary œdema. Exploration alone was done in 24 patients with four deaths.

The subclavian artery has been ligated in about 555 patients without difficulty. The ligature is usually placed just distal to the branching of the vessel. A block of the sympathetic nerves has never been necessary. The arm and hand supplied by this vessel are customarily cooler for several days after the operation, and no pulse is felt for a considerably longer time. The patients often state that the arm feels weaker for several days, but there has never been a significant interference with function.

Most of the patients who survive the operation are improved. This improvement varies in degree from that of patients who are considerably restricted to those who have no limitation of their activities. In patients with a satisfactory result the colour improves rapidly following the completion of the anastomosis. The oxygen saturation rises, the polycythemia diminishes and may disappear, and the clubbing of the digits regresses. The pulse pressure increases, and there is a soft continuous murmur audible over the chest. The most gratifying alteration is the increased capacity for exertion. Some patients who previously were limited to a few steps can now walk miles, and many are able to exercise normally and equal other children of their age in activity. The point must be stressed, however, that the interval since operation is as yet too short to allow an accurate evaluation of the final results.

Summary

The indications for operation and the operative procedures performed upon 610 patients with congenital heart disease of the cyanotic type are described. The anomalies of blood vessels which have been encountered are enumerated, and a brief summary of the results is given.

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ELECTION OF PRESIDENT AND VICE-PRESIDENTS—8th JULY, 1948

Lord Webb-Johnson was re-elected President for the eighth year. Mr. L. E. C. Norbury was re-elected Vice-President, Mr. Zachary Cope was elected Vice-President for the ensuing year.